Case Report

Epithelioid Hemangioma of the Thoracic Spine: A Case Report and Review of the Literature

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Context: Osseous epithelioid hemangioma is uncommon, and reports of epithelioid hemangiomas of the spine are especially rare.
Study Design: Case report.
Findings: A 43-year-old male was referred to our department with progressive gait disturbance. CT scans showed a lucent mass in the vertebral body at the T3 level. MRI of the thoracic spine showed a strongly enhanced mass compressing the spinal cord. The patient underwent laminectomy from T2 to T4, debulking of the tumor, and posterior fusion from T1 to T5. After the operation, the patient's neurological status improved significantly, and he was able walk without assistance. Histological examination determined that the tumor was an epithelioid hemangioma. The patient was treated with 40 Gy radiation for local control of the tumor. The patient could walk without difficulty 12 months after the surgery.
Conclusion: This is a rare example of an epithelioid hemangioma that developed in the thoracic spine and compressed the spinal cord, and was treated successfully.

Keywords: Epithelioid hemangioma, thoracic spine, spinal compression, spinal instrumentation, spinal tumor, benign tumor, surgical treatment, radiation

Introduction

Epithelioid hemangioma (EH), a rare mesenchymal tumor of vascular origin, is benign but locally aggressive.[1,2] EH usually occurs as a reddish-brown mass forming in the skin of the head, neck, or extremities, and is mostly found in young and middle-aged adults. EH was first recognized as a distinct entity in 1983, and has been called angiolymphoid hyperplasia with eosinophilia, inflammatory angiomatous nodule, and histiocytoid hemangioma. The World Health Organization defines EH as a distinct tumor type[3] that is sometimes difficult to distinguish from endothelioid hemangioendothelioma, a tumor of intermediate malignancy. When EH occurs in osseous tissues, it is found most frequently in the long tubular bones of the extremities,[4] followed by the short tubular bones of the distal lower extremities and flat bones.[5] The reported prevalence of EH of the spine is 6–16% of osseous EH.[4,6] Most EH presents without neurological symptoms, and primary EH of the spine with neurological deficits appears to be extremely rare.[7,8] Here we present a very rare case of EH compressing the spinal cord.

Case presentation

A 43-year-old male was referred to our department with a 2-week history of back pain and progressive gait disturbance. The patient began to experience numbness starting below the epigastric fossa and radiating down both legs to the toes. He had no bladder or rectal disturbance. He had normal strength in his arms, but a weakness of MMT grade 4 in his legs. He had difficulty standing or walking without support, and could not ascend stairs without the assistance of a handrail. Hyperactive deep tendon reflexes and Babinski's sign were present on both sides.

A serial radiographic survey of the thoracic spine was unremarkable except for mild degenerative changes,

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including vertebral osteophytes. A computed tomographic (CT) scan of the thoracic spine showed a multilocular and expanding lytic lesion at T3 in the vertebral body, pedicles, and lateral processes (Fig. 1). Magnetic resonance imaging (MRI) of the thoracic spine demonstrated a diffusely enhanced spinal tumor with low intensity on T1-weighted MRI and hyperintensity on T2-weighted MRI. The tumor extended into the spinal canal and was compressing the spinal cord at the T3 level (Fig. 2).

We treated the tumor by embolizing two tumor vessels intravascularly, followed by a laminectomy from T2 to T4, intralesional tumor debulking, and posterior fusion from T1 to T5 using a pedicle-screw system. Intraoperatively, we found that the tumor involved the T3 vertebral body and the transverse processes on both sides. The tumor was brittle and bled massively during resection. We controlled the bleeding with bone wax and resected the tumor in a piecemeal fashion using an air drill with a diamond burr and a Cavitron Ultrasonic Surgical Aspirator (Valleylab, Boulder, CO, USA). The operation took 240 minutes, and the estimated blood loss was 1700 ml.

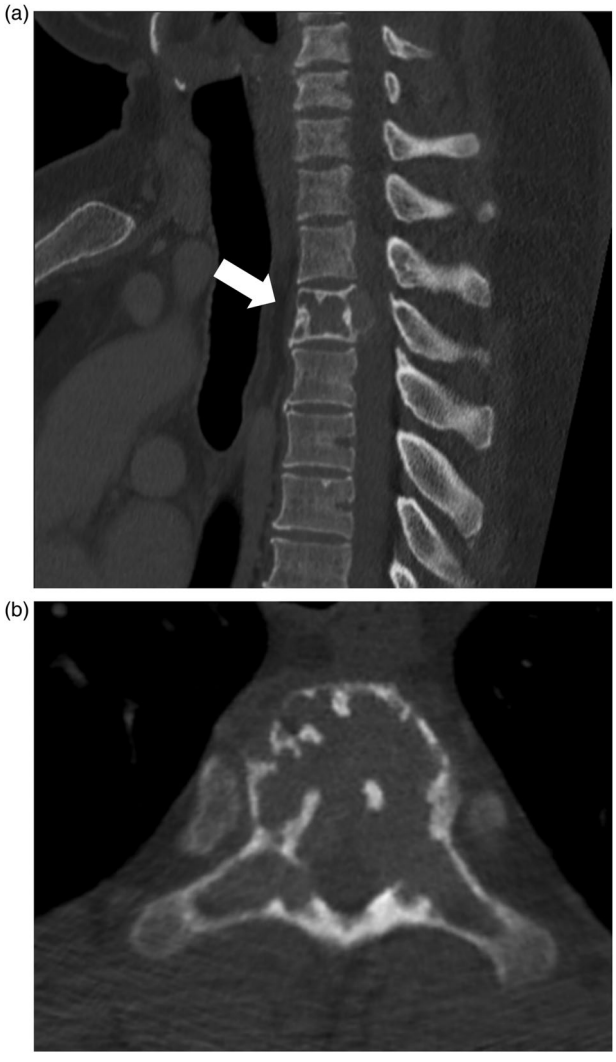

Figure 1. (A) CT sagittal view of the thoracic spine, showing a spinal tumor at the T3 vertebral body expanding into to the spinal canal (arrow). (B) CT axial view of the thoracic spine, showing a multilocular and expanding lytic lesion in the vertebral body and bilateral transverse process.

The specimen submitted for pathological examination consisted of fragmented pieces of bony tissue. Histologically, the bone marrow was occupied by loose fibrous tissue containing many capillary-sized vessels lined by epithelioid endothelial cells (Fig. 3). Some vessels had an open lumen; in others, the lumen was obliterated by the proliferation of epithelioid endothelial cells. Immunohistochemically, the epithelioid endothelial cells were positive for the endothelial markers CD31 and factor VIII–related antigen, confirming the endothelial nature of the cells. Another endothelial maker, CD34 was negative. There was no prominent nuclear atypia or abundance of mitoses. The Ki-67 labeling index was less than 1%, indicating very low proliferative activity. Hemangioepithelioid endothelioma was excluded because the tissue lacked the characteristics of poorly formed blood vessels or myxoid to hyalinized stroma.

The patient's neurological status improved significantly after the surgery. He was treated with 40 Gy radiation to prevent local recurrence. One year after surgery, the patient was able to walk without a cane or walker, and MRI of the thoracic spine showed no local tumor recurrence. There was no sign of systemic metastasis.

Discussion

The malignant potential of EH remains controversial. While EH is generally considered benign, Nielsen *et al.* recently demonstrated that osseous EH is an aggressive neoplasm with rates of local recurrence and distinct metastasis of 11% and 2.7%, respectively,[4] and thus advocated classifying osseous EH as a neoplasm with low but definite malignant potential.

Previous reports indicate that EH in the spine arises mostly from within the vertebral body. However, transverse processes can also host EH. In the present case, the tumor located at the bilateral transverse processes and the vertebral body.Although the patient only noted back pain and difficulty walking for a period of two weeks, the tumor might have been gradually expanding for a longer period. In general, EH is lucent in plain radiographs and CT scans. Typical EH characteristics on MRI are hypo- to isointense on T1-weighted images, hyperintense on T2-weighted images, and

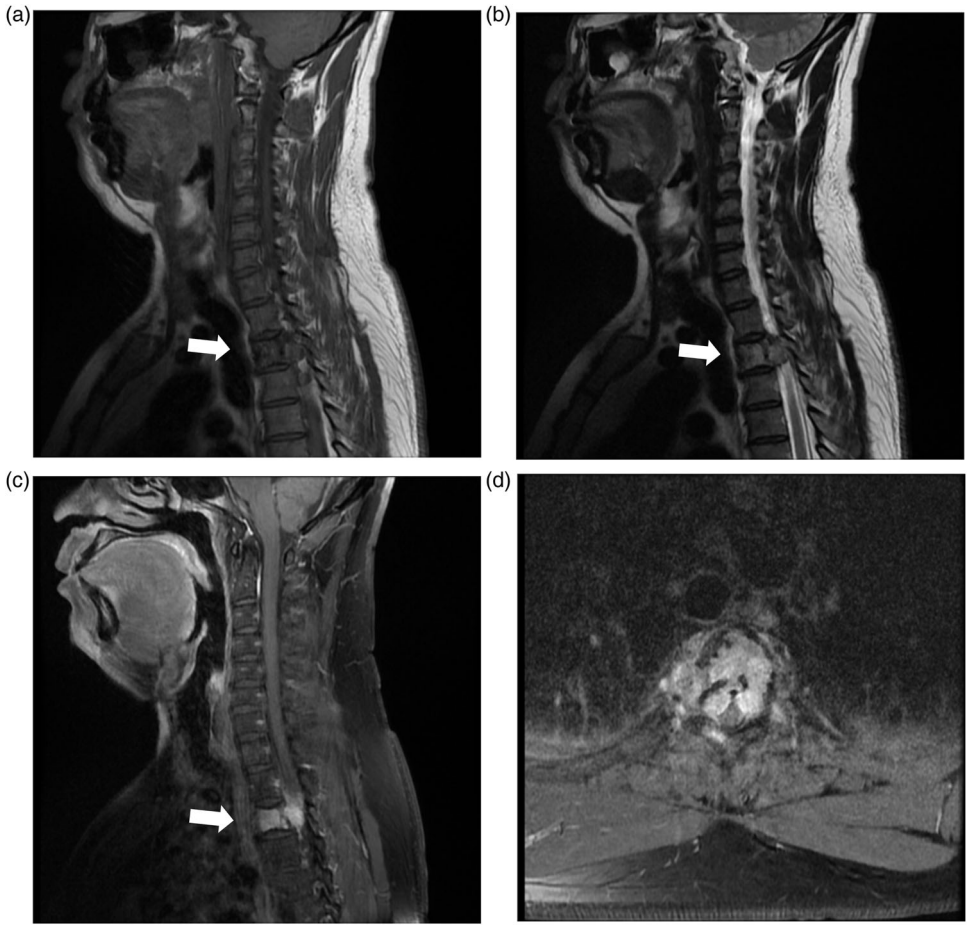

Figure 2. MRI of the thoracic spine shows a T1-weighted hypointense, T2-weighted hyperintense, and avid enhanced mass at the T3 level (arrow). (A) Sagittal T1-weighted image, (B) sagittal T2-weighted image, (C) sagittal enhanced image, and (D) axial enhanced image at the T3 level.

markedly enhanced by gadolinium.[9] These were precisely the characteristics observed in our present case.

Only 21 cases of EH of the spine are reported in the English literature, including the present case.[6–11] We excluded eight cases reported by Nielsen *et al.*[4] because of insufficient detail, and reviewed the remaining 13 reported cases of EH of the spine (Table 1). Although EH typically occurs during early to mid-adult life (20–40 years of age),[1] patients with EH of the spine ranged from 20 to 65 years (mean 44±13), and thus were older than patients with EH of the soft tissue or bones. Although EH is more frequent in females,[1] EH of the spine was more prevalent in males (n=9, 69%). Although most cases of EH of the spine involved the thoracic spine, one case arose in the cervical spine and another in the lumbar spine.[8,11] Among the 13 cases, four patients (31%) developed EH in two adjacent levels of the spine. Although local pain was present in all cases (100%), neurological symptoms were present in only six cases (46%).

The differential diagnosis of EH in the spine was shown in table 2. Radiographically, osteolytic lesion is observed in aneurysmal bone cyst, giant cell tumor, chondrosarcoma, hemangioma, and EH. These bone tumors usually occur in middle-age. Aneurysmal bone cyst and giant cell tumor appear commonly in relatively younger age than the other tumor. Thoracic spine is the frequent location in aneurysmal bone cyst, chondrosarcoma and EH. On MRI, hyperintence lesions were seen on T2WI in these tumors except for giant cell tumors.

Except for one patient who underwent biopsy only (8%) without further treatment, the patients were treated by surgery with total resection (n=4, 31%) or partial resection (n=8, 62%). Additional radiation therapy was given in eight cases (62%). In 2016, Zhou *et al.* reported that EH of the cervical spine recurred locally four years after partial resection.[11] However, no deaths related to EH have been reported (6). Whether en bloc or piecemeal resection is optimal for treating spinal EH is controversial. Although the malignant potential of EH of the spine makes total resection desirable, radical resection might result in massive bleeding or neurological deterioration. According to

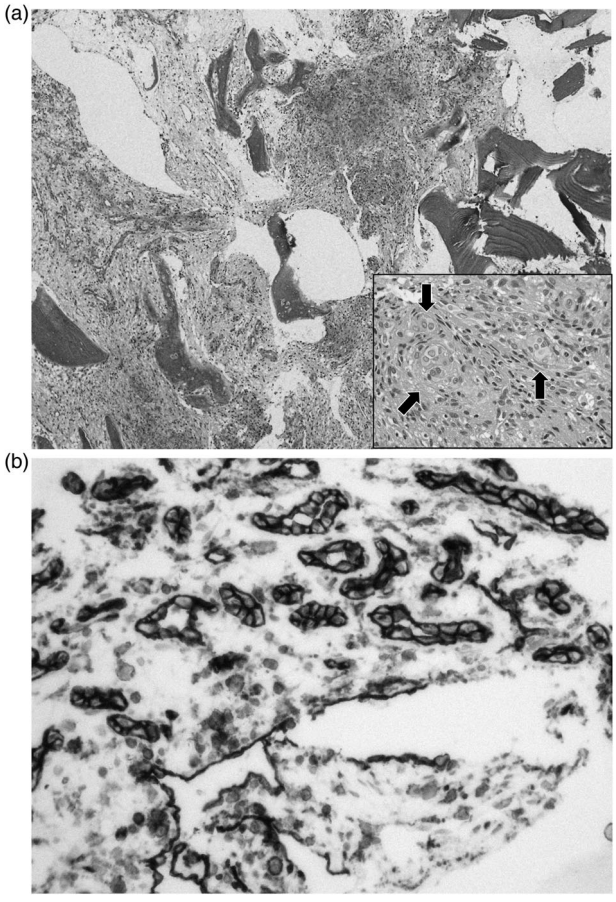

Figure 3. Histologic and immunohistochemical findings. (A) Bone tissue resected from the T3 vertebral body had fibrous tissue in the marrow spaces with many capillary-sized blood vessels. Inset: Magnified view of the blood vessels lined by epithelioid endothelial cells (arrows) (HE stain). (B) Immunohistochemistry showed epithelioid endothelial cells positive for CD31.

our literature review, no case with systemic metastasis of EH primarily occurred at the spine had been reported. As WHO classification 2, we considered EH in the spine have an intermediate malignancy. Our review of the literatures suggested that total resections were not always necessary for spinal EH, since most tumors follow a benign course except for one case (7.7%) with local recurrence. In the present case, in spite of preoperative intravascular embolization, we could not totally resect the tumor due to intraoperative massive bleeding. Therefore, an additional radiotherapy was applied to avoid local recurrence.

Our case illustrates the clinical and radiographic characteristics of spinal EH, and suggests that piecemeal resection of the tumor followed by radiotherapy can be an acceptable treatment choice, although more cases will need to be studied before drawing conclusions about the optimal treatment.

Table 1. Summary of Previously Reported Epithelioid Hemangiomas in the Spine

Author	Year	Sex	Age	Level	Neurological Symptoms	Treatment	Total resection	Radiation	Prognosis
Ling7	2001	F	32	T7	No	S	Yes	No	AW 48 months
Errani6	2012	M	50	T2, T3	ND	S	No	Yes	DOO 16 months
Boyaci8	2013	F	49	L3	Yes	S	Yes	No	AW 25 months
		M	49	T4	Yes	S	No	Yes	AW 26 months
		M	58	T6	No	S	No	Yes	AW 22 months
		M	20	T8	No	S	Yes	No	AW 6 months
		M	37	L1	No	None (biopsied) S	No	No	AW 87 months
		M	29	T4	Yes	S	Yes	Yes	AW 115 months
O'Shea9	2014	M	49	T6, T7	Yes	S	No	Yes	ND
		M	50	T3	No	S	No	Yes	ND
Weaver10	2015	F	65	T7, T8	Yes	S	No	No	ND
Zhou11	2016	F	43	C5, C6	ND	S	No	Yes	LR 24 months
Present case	2017	M	43	T5	Yes	S	No	Yes	AW 12 months

AW: alive and well; DOO: died of other causes; LR: local recurrence; ND: not described; S: surgery. .

Table 2. Differential diagnosis of epithelioid hemangioma of spine.

	Age Sex	Location	CT images features	MRI features	Key facts
Aneurysmal bone cyst[12,13]	< 30 y F > M	Lumbar spine Posterior element or pedicle	Osteolytic lesion, cortical eggshell	Heterogeneous lesions Hypointense and hyperintence on T1 – and T2WI	Local curettage is the treatment option, Local recurrence may occur
Giant cell tumor[14–16]	20–40 y F>M	Sacrum Vertebral body	"Soap bubble" expansive osteolytic lesion	Hypointense on T2WI.	Aggressive resection is needed. Local recurrence and sometime metastasize
Chondrosarcoma[17,18]	50–60 y M >F	Thoracic spine	Expansive osteolytic lesions with diffuse, mottled "ring and arc " calcification	Hypointense and hyperintence on T1 – and T2WI	Complete en-bloc surgical resection is needed.
Hemangioma[19,20]	Incidence increases with age F > M	Vertebral body > posterior element	"Honeycomb" appearance and "polka dot" appearance on axial CT	Hyperintense signal on both T1- and T2WI	In aggressive type, tumor may extend epidurally and cause cord compression
Epithelioid hemangioma[4,6–11,21]	20–65 years M > F	Thoracic spine Transverse process	Multilocular and expanding lytic lesion	Isointence on T1WI, hyperintence on T2WI	Local recurrence and may occur.

Our patient consented to the publication of his data and images in this case report.

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